Tetralogy of Fallot With Diminutive Pulmonary Arteries: Preoperative Pulmonary Valve Dilation and Transcatheter Rehabilitation of Pulmonary Arteries

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Objectives. This study sought to determine the results of a novel transcatheter management approach in tetralogy of Fallot with diminutive pulmonary arteries.

Background. Tetralogy of Fallot with diminutive pulmonary arteries and severe pulmonary stenosis is rare and resembles tetralogy of Fallot with pulmonary atresia: There is a high incidence of aortopulmonary collateral channels, arteriovenous anomalies, stenoses and need for multiple operations. Because a combined catheter-surgery approach facilitates repair in these patients, such an approach may benefit those with diminutive pulmonary arteries and pulmonary stenosis.

Methods. Clinical, catheterization and surgical data were studied retrospectively for 10 such patients undergoing preoperative pulmonary valve balloon dilation, among other transcatheter interventions, from January 1989 to January 1995.

Results. Initially, the Nakata index ranged from 20 to 98 mm²/m² (mean 67 ± 28 mm²/m²). The pulmonary valve was first balloon dilated (mean balloon/annulus 1.5 ± 0.3), and the mean initial valve annulus Z score (−4.0 ± 1) increased to −3.3 ± 1.1 (p < 0.01). Other interventions included branch pulmonary artery balloon dilation (7 patients, 23 vessels) and coil embolization of aortopulmonary collateral channels (8 patients, 31 collateral channels). At preoperative catheterization, the annulus pulmonary annulus Z score was −3.1 ± 0.7, and the aortic arch index increased to 143 ± 84 mm²/m² (p < 0.03). All patients underwent complete surgical repair successfully. At a mean follow-up period of 2.6 ± 0.2 years, right ventricular pressure was <70% systemic in all patients and <50% systemic in seven.

Conclusions. In patients with tetralogy of Fallot, severe pulmonary stenosis and diminutive pulmonary arteries, initial pulmonary valve balloon dilation increases the annulus Z score and antegrade pulmonary blood flow and facilitates simultaneous coiling of aortopulmonary collateral channels and access for branch pulmonary artery dilation, all of which results in pulmonary artery growth, simplifying surgical management.

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There is a wide spectrum of disease in tetralogy of Fallot, ranging from mild forms of infundibular obstruction and pulmonary valve stenosis to pulmonary atresia with hypoplastic or even absent pulmonary arteries (1–5). Most patients with tetralogy of Fallot can undergo complete primary repair during infancy (6–8) without occcluding a pulmonary artery (1–5). In contrast, patients with rare, severe forms with diminutive pulmonary arteries in whom the pulmonary blood flow is derived primarily from collateral channels usually need multiple interventions before complete repair (9,10) because the central pulmonary arteries may be inadequate to carry a full cardiac output after ventricular septal defect closure. Some have even advocated lifelong medical management for these patients (11).

Since 1984, we have managed most patients with tetralogy of Fallot, pulmonary atresia and diminutive pulmonary arteries with an initial right ventricle-to-pulmonary artery graft, followed by balloon dilation of pulmonary artery stenoses and coil embolization of aortopulmonary collateral channels and, later, surgical closure of the ventricular septal defect in addition to repair of any remaining central obstructions (7.9). Such a management approach, combining interventional catheterization and operation, seems to enhance the chances of achieving a satisfactory repair (9).

On the basis of this favorable experience, we dilated the pulmonary valve in a group of 10 patients with severe tetralogy of Fallot, diminutive pulmonary arteries or multiple aortopulmonary collateral channels, thought to be unsuitable for primary complete surgical repair on presentation. In these patients, pulmonary valve dilation was an initial step for transcatheter pulmonary artery "rehabilitation," followed by coil embolization of aortopulmonary collateral channels, dilatation...
tion of distal pulmonary artery stenosis and subsequent complete surgical repair.

Methods

Study group. All unoperated patients with tetralogy of Fallot who underwent any preoperative catheterization from January 1984 to January 1995 were identified from the Department of Cardiology data base. Patients with angiographic diagnosis of pulmonary artery hypoplasia or branch pulmonary artery stenosis were reviewed. Of those, patients with diminutive pulmonary arteries (Nakata index < 100 mm²/m²) who had undergone pulmonary valve balloon dilation as initial intervention were selected and constitute the study group. Because most patients with usual tetralogy of Fallot underwent primary repair after a diagnostic echocardiogram without the need for a previous catheterization, such patients were not included in this review. Patients with any previous surgical intervention (such as Blalock-Taussig shunts) or with diagnoses of pulmonary atresia, absent pulmonary valve syndrome, associated other major cardiac malformations (such as endocardial cushion defects, abnormalities in the ventricular loop, transposition of the great arteries, heterotaxia syndromes or hypoplastic ventricles) were excluded.

Medical records, echocardiograms and angiographic and hemodynamic data were reviewed in all patients from the time of presentation to the most recent follow-up date. Approval for chart review was obtained from the Institutional Review Board.

Cardiac catheterization. At initial catheterization, after routine right- and left-sided hemodynamic analysis, patients had determination of 1) pulmonary valve annulus Z score; 2) right and left pulmonary artery Z scores; 3) Nakata index; 4) McGoon ratio (12); 5) presence of aortopulmonary collaterals or patent ductus arteriosus; 6) oxygen saturation before and after pulmonary valve balloon dilation; and 7) other transcatheter interventions. The pulmonary artery size was determined by measuring the right and left pulmonary artery diameters immediately proximal to their first branch points at the initial angiogram. Magnification errors were corrected by relating vessel size to the known diameter of the angiographic catheter. The Nakata index was reported for each patient as the sum of the cross-sectional areas of both pulmonary arteries indexed to the body surface area (13). The normal Nakata index is 330 ± 30 mm²/m². Pulmonary arteries with Nakata index less than 100 mm²/m² were considered diminutive. The Z values obtained were based on the reported definition of Z = observed dimension minus mean normal dimension/standard deviation (14). Main pulmonary artery pressure and oxygen saturation were determined before and after dilation in some patients, with the exception of those with such severe stenosis that no pressure-recording catheter could be advanced across the valve before dilation. The presence of other sources of pulmonary blood flow by aortopulmonary collaterals or a patent ductus was determined by aortography or selective collateral injection.

At later preoperative catheterization, patients had repeat determination of 1) pulmonary valve annulus Z score; 2) right and left pulmonary artery Z scores; and 3) Nakata index. The initial measurements at the first catheterization before any intervention were compared with those immediately following transcatheter intervention and those at a preoperative catheterization.

Transcatheter interventions performed at preoperative catheterizations included pulmonary valve balloon dilation as previously described (15,16), branch pulmonary artery balloon dilation (17–20), coil embolization of aortopulmonary collaterals (21) in most patients and stent placement in the right ventricular outflow tract in one patient.

Parental informed consent was obtained before every procedure performed.

Operation. Operative notes and immediate and late postoperative results were reviewed. The surgical technique used for tetalogy of Fallot repair has been well described previously (22).

Follow-up. Follow-up information was obtained from hospital records and from referring physicians. At the latest follow-up, the results of electrocardiograms, echocardiograms, oximetry, available catheterizations and symptoms were reviewed. Right ventricular pressure by noninvasive (echocardiogram and Doppler) or invasive methods (cardiac catheterization) was determined in all patients at follow-up.

Statistical analysis. Results obtained are expressed as mean value ± SD. Pulmonary valve annulus diameter and right and left pulmonary arteries were expressed as Z scores (number of standard deviations from the mean normal for body surface area) (14,23). Initial results, immediate results after balloon dilation and late results at preoperative follow-up catheterization were compared and tested with a paired t test.

Results

Patient characteristics. Ten patients with diminutive pulmonary arteries (17 days to 12 years old, mean age 1.9 ± 3.7) underwent pulmonary valve balloon dilation as initial preoperative transcatheter intervention since June 1989. There were four boys and six girls. Age, weight and oxygen saturation at pulmonary valve balloon dilation, age at surgical correction and follow-up period after operation are summarized in Table 1.

Among associated anomalies were right aortic arch in four patients, aberrant right subclavian artery in one and restrictive ventricular septal defect with suprasystemic preoperative right ventricular pressures in three patients. Partial Di George syndrome was diagnosed in one patient. One patient had a persistent left superior vena cava to the coronary sinus.

Cardiac catheterization. Pulmonary artery characteristics. All patients had more than one catheterization, with an average total number of 2.6 per patient. All patients had diminutive pulmonary arteries (Fig. 1 and 2) with an average Nakata index of 67 ± 28 mm²/m², ranging from 20 to
### Table 1. Patient Characteristics

<table>
<thead>
<tr>
<th>Pt No.</th>
<th>Age at Operation (yr)</th>
<th>Gender</th>
<th>Weight (kg)</th>
<th>Oxygen Sat. (%)</th>
<th>Age at Follow-Up (yr)</th>
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<tr>
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<td>2.6</td>
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<tr>
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<tr>
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<td>66</td>
<td>1.5</td>
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<td>76</td>
<td>0.5</td>
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</tbody>
</table>

Average ± SD: 1.9 ± 3.7, 8.9 ± 7, 78.7 ± 8, 25 ± 3.6, 2 ± 2.1

BD = pulmonary valve balloon dilation; Oxygen Sat. (%) = percent oxygen saturation at rest in room air, before any intervention; Pt = patient.

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**Figure 1.** A. Right pulmonary artery angiogram demonstrates diminutive pulmonary artery. Note that contrast in right lower lobe is being washed by competing collateral flow. B. Selective angiogram of aortopulmonary collateral flow to left lower lobe demonstrates its dual supply with filling of diminutive central pulmonary arteries (arrow).

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**Figure 2.** A. Before interventions. Pulmonary artery angiogram at initial catheterization demonstrates hypoplastic pulmonary arteries with severe stenosis of the proximal left pulmonary artery. Note diminished filling of left lung and right lower lobe as contrast is washed by aortopulmonary collateral flow. The pulmonary valve annulus is markedly hypoplastic with thickened leaflet (arrow). B. After interventions. Pulmonary artery angiogram of the same patient as A, after all transcatheter interventions and operation, demonstrates marked improvement in vessel dimensions and flow. Note that the aortopulmonary collateral channels have been coil embolized.
Table 2. Angiographic Characteristics and Summary of Transcatheter Interventions

<table>
<thead>
<tr>
<th>Pt No.</th>
<th>Nakata Index (mm²/m²)</th>
<th>McGoon Ratio</th>
<th>MAPCs</th>
<th>Branch PA Stenosis</th>
<th>Coil MAPCs</th>
<th>Other Interventions</th>
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<td>8</td>
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<tr>
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<td>0.8</td>
<td>Yes</td>
<td>Yes</td>
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</table>

Average: 67 ± 28 mm²/m²

ASD = atrial septal defect; BD = balloon dilation; MAPCs = multiple aortopulmonary collateral channels; McGoon Ratio = Right pulmonary artery (PA) diameter + Left pulmonary artery diameter/Descending aorta diameter; Pt = patient; RVOT = right ventricular outflow tract.

98 mm²/m² (Table 2). The right and left pulmonary artery Z scores ranged from −4.8 to −2.0 (mean −2.8 ± 0.9) and −4.5 to −0.5 (mean −2.2 ± 1), respectively. The McGoon ratio was below 1.5 for all patients, ranging from 0.6 to 1.2 (average 0.8 ± 0.2). Eight patients had multiple aortopulmonary collaterals. All lung segments supplied by aortopulmonary collaterals had dual supply by the native central pulmonary arteries as well (Fig. 1).

Pulmonary valve balloon dilation. Before balloon dilation, the pulmonary valve annulus Z score ranged from −5.8 to −3.1 (average −4.0 ± 1). Pulmonary valve dilation using a balloon/annulus ratio of 1.5 ± 0.3 resulted in a small immediate increase in the valve annulus size (Fig. 3). In seven patients there was an increase in initial diameter of at least 25%. Immediately following dilation, the average pulmonary valve annulus Z score increased from −4.0 to −3.3 (p < 0.001). The average number of valve dilations per patient was 3, ranging from 1 to 10.

There was also documentation of a small increase in the oxygen saturation ≥4% immediately following the procedure in five patients. However, because of simultaneous coil embolization of aortopulmonary collaterals, the change in systemic oxygen saturation was not consistent. In eight patients with adequate measurement of the main pulmonary artery pressure both before and after balloon dilation, an increase from a mean of 19 ± 7 to 30 ± 12 mm Hg (p < 0.02) was observed.

Coil embolization of aortopulmonary collateral channels. Eight patients underwent coilng of aortopulmonary collaterals, most of which were occluded during the same initial catheterization (six patients), although two patients had them coiled at subsequent catheterizations. The total number of aortopulmonary collaterals coil embolized was 31. Thirty were successfully occluded, with no flow or trivial residual flow by selective angiography following the procedure. One patient had a large collateral ligated via video-assisted thoracoscopic surgery as a result of unsuccessful coil occlusion.

Balloon dilation of branch pulmonary artery stenoses. Seven patients underwent branch pulmonary artery balloon dilation, with a total of 22 vessels dilated, including segmental, lobar and main pulmonary artery branches. The mean average vessel diameter increased significantly with balloon dilation (p < 0.001). The diameter increased more than 30% of the initial size in 15 of the 22 vessels dilated. All but two vessels had improvement in the diameter and angiographic flow with balloon dilation. The average diameter increase was 76 ± 88%.

Other interventions. One patient with multiple very large aortopulmonary collaterals and diminutive pulmonary arteries underwent placement of a stent in the right ventricular outflow tract at a second preoperative catheterization in an attempt to improve antegrade pulmonary blood flow and allow coil occlusion of the large collaterals.

Preoperative catheterization. At preoperative catheterization, the Nakata index improved from 67 ± 28 mm²/m² at presentation to 143 ± 84 mm²/m² preoperatively (p < 0.03)
Fig. 4. Comparison of Nakata index before balloon dilation (BD) and any interventions with values obtained at preoperative catheterization in seven patients. Note that there is a significant improvement in Nakata index in all but one patient before operation.

(Fig. 4). Similarly, the right pulmonary artery Z scores changed from $-2.8 \pm 0.8$ to $-0.7 \pm 2$ ($p < 0.05$), and the left pulmonary artery Z scores increased from $-2.2 \pm 1.3$ to $-1.5 \pm 1.9$ ($p < 0.07$) (Fig. 5, Table 3). The average pulmonary valve annulus Z score, which had increased from $-4 \pm 0.1$ before dilation to $-3.3 \pm 1.1$ immediately after dilation, was $-3.1 \pm 0.7$ preoperatively (Fig. 3).

Complications. There were no deaths. No tetralogy of Fallot cyanotic spells were observed during or after interventional catheterization. One patient, in whom a large aortopulmonary collateral could not be closed in the catheterization laboratory, had transient congestive heart failure and pulmonary edema requiring an additional 10 days in the intensive care unit with positive-pressure mechanical ventilation for 5 days. Because of collateral "steal" of descending aorta blood flow, this same patient had necrotizing enterocolitis, managed medically. During an attempt at coil embolization of the aforementioned large collateral, a coil was dislodged in the pulmonary artery distal collateral end. It was successfully retrieved, but damage to the vessel wall (contrast staining) without extravasation was observed by angiography. Subsequently, this large collateral was successfully ligated by videoassisted thoracotomy. Another patient developed significant lobar pulmonary edema secondary to branch pulmonary artery balloon dilation and also required transient mechanical ventilation. Occlusion of the femoral arteries bilaterally was a complication in one patient with multiple aortopulmonary collaterals, which were coil embolized via arterial access. One patient had a transient pulse loss, and three had unilateral femoral vein occlusion diagnosed at a late catheterization.

Surgical management. Nine of 10 patients underwent complete single-stage surgical correction at an average of 8.5 ± 0.6 months (range 1 to 23 months) following the initial catheterization. One patient had a staged surgical repair with initial right ventricle-to-pulmonary artery homograft followed by ventricular septal defect closure 7 months later. The corrective surgical procedure involved ventricular septal defect closure in all and transannular right ventricular outflow tract patch in eight patients. One of these eight patients had augmentation of the outflow tract patch with use of a unicusp aortic homograft. Two patients had placement of a right ventricle-to-pulmonary artery homograft.

Figure 5. Left pulmonary artery (LPA) and right pulmonary artery (RPA) Z scores before any interventions and at preoperative follow-up catheterization for each patient. BD = balloon dilation.

Outcome and follow-up. All patients underwent complete operation at an average of 6 months after the first interventional catheterization. The mean follow-up period after operation was 2 years, ranging from 1 month to 4.9 years. At follow-up, the right ventricular pressure, estimated by echocardiogram (10 patients) or catheterization (6 patients), or both, was ≤ 50% systemic in 7 patients and between 50% and 70% in 3 patients. 2 of whom had right ventricle-to-pulmonary artery homografts placed, which were becoming relatively small with time. No patient had a right ventricular pressure higher than 70% systemic at follow-up. The average oxygen saturation at follow-up was 96%.

During follow-up, two patients had a residual atrial septal defect closed with a clampsell device in the cardiac catheterization laboratory without complications 4 and 10 months following complete tetralogy of Fallot repair, respectively.

Discussion

We report the results of a novel management approach for patients with severe tetralogy of Fallot and pulmonary stenosis who are thought to be unsuitable for complete surgical repair in one stage, involving use of transcatheter techniques.

Management of patients with tetralogy of Fallot. Tetralogy with diminutive pulmonary arteries and multiple aortopulmonary collaterals, in the absence of pulmonary atresia, is rare (2,3). Such patients share common characteristics with patients

| Table 3. Average Values of Pulmonary Valve Annulus, Right and Left Pulmonary Artery Z Scores and Nakata Index Before Catheter Intervention and at Preoperative Catheterization |
|---|---|---|---|
| PVA Z score | $-4.0 \pm 1$ | $-3.1 \pm 0.7$ | < 0.01 |
| RPA Z score | $-2.8 \pm 0.8$ | $-0.7 \pm 2.3$ | < 0.05 |
| LPA Z score | $-2.2 \pm 1.5$ | $-1.5 \pm 1.9$ | < 0.07 |
| NI | $167 \pm 82$ | $143 \pm 84$ | < 0.03 |

*Values at preoperative follow-up catheterization after transcatheter interventions had been performed. LPA = left pulmonary artery; NI = Nakata index (mm²/m²); PVA = pulmonary valve annulus; RPA = right pulmonary artery.
with tetralogy and pulmonary atresia and represent a group with a higher degree of severity, with features that overlap tetralogy with pulmonary atresia and tetralogy with pulmonary stenosis. Patients in whom the pulmonary arteries are thought to be too small to undergo complete correction generally undergo initial palliative surgical interventions. Even though surgical shunts have been shown to increase pulmonary artery growth (24,25) in patients with small pulmonary arteries, the effectiveness for diminutive pulmonary arteries (i.e., less than 100 mm²/m²) remains unproven. In addition, various studies, including postmortem studies, have demonstrated markedly abnormal pulmonary artery development in patients with shunts (26), including pulmonary artery distortion, inhomogeneous perfusion and multiple potential postoperative complications (27-30). In patients with hypoplasia or branch pulmonary artery stenosis, an aortopulmonary shunt offers very limited possibilities for subsequent transcatheter balloon dilation. Since 1984, we have opted to surgically establish right pulmonary artery growth and both anatomic and physiologic preoperative conditions. Most patients with tetralogy of Fallot do not need palliative procedures or even preoperative catheter intervention to study the factors predictive of success of this management approach.

Conclusions. Initial pulmonary valve balloon dilation in patients with tetralogy of Fallot, severe pulmonary stenosis and diminutive pulmonary arteries increases the annulus Z score and anterograde pulmonary blood flow, allowing simultaneous coiling of aortopulmonary collaterals and easier access for branch pulmonary artery dilation, all of which improve pulmonary artery growth and both anatomic and physiologic preoperative conditions. Most patients with tetralogy of Fallot do not need palliative procedures or even preoperative catheterization. In contrast, patients with diminutive pulmonary arteries seem to benefit from an initial preoperative transcatheter dilation, although the small size of the series, the multiplicity of interventions performed and the lack of adequate control group make this conclusion tentative.

References


